

Case Report

Retroperitoneal fibrosis: a rare cause of recurring abdominal pain

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Retroperitoneal fibrosis (RPF) is a rare condition of unclear aetiology. It is believed to be immune-related. About two-thirds of cases are thought to be idiopathic.¹ We present a case of idiopathic RPF in a 37-year old male with recurring abdominal pain over a five-month period associated with features of ischaemic colitis and bilateral hydroceles. An initial CT scan of the abdomen showed a significant peri-aortic soft tissue mass. The inferior mesenteric artery (IMA) was noted to pass through the mass and to be compressed by this mass. A subsequent CT-guided biopsy confirmed retroperitoneal fibrosis. He was successfully treated with steroids only with resolution of his symptoms and radiological features.

To our knowledge no case of idiopathic RPF, presenting with features of ischaemic colitis and bilateral hydroceles, has been reported in the UK.

CASE REPORT Mr RS, a 37-year old male, first presented to the Trust via his General Practitioner with a three-week history of intermittent sharp flank and left iliac fossa (LIF) pains radiating to his left testicle. He had no significant past medical history.

Examination revealed some tenderness in the left loin and left iliac fossa areas with no other remarkable findings. CRP and ESR were significantly elevated but other baseline laboratory tests were normal. He had an Ultrasound Scan (USS) of the abdomen which was essentially normal. His symptoms settled with conservative management and he was discharged a few days later.

He was however readmitted a week later with a recurrence of the LIF pain but this time associated with bloody diarrhoea. Repeat investigations including *Clostridium difficile* were all normal except CRP, which was again significantly elevated. He

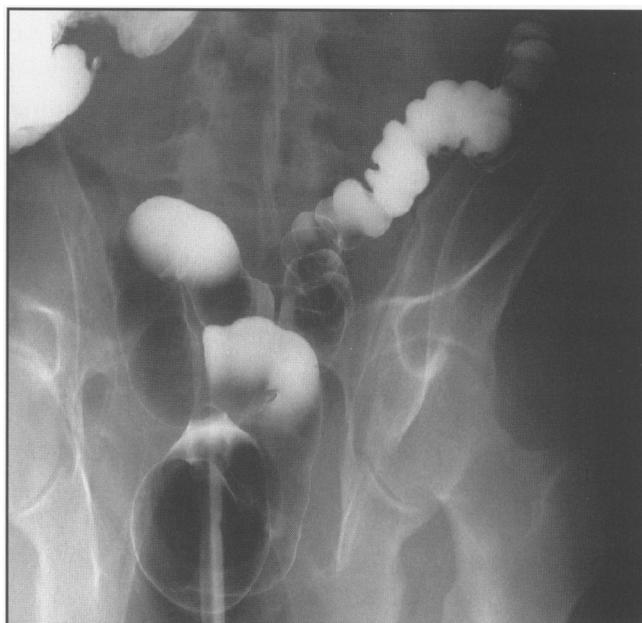


Fig 1. Initial Barium Enema: normal.

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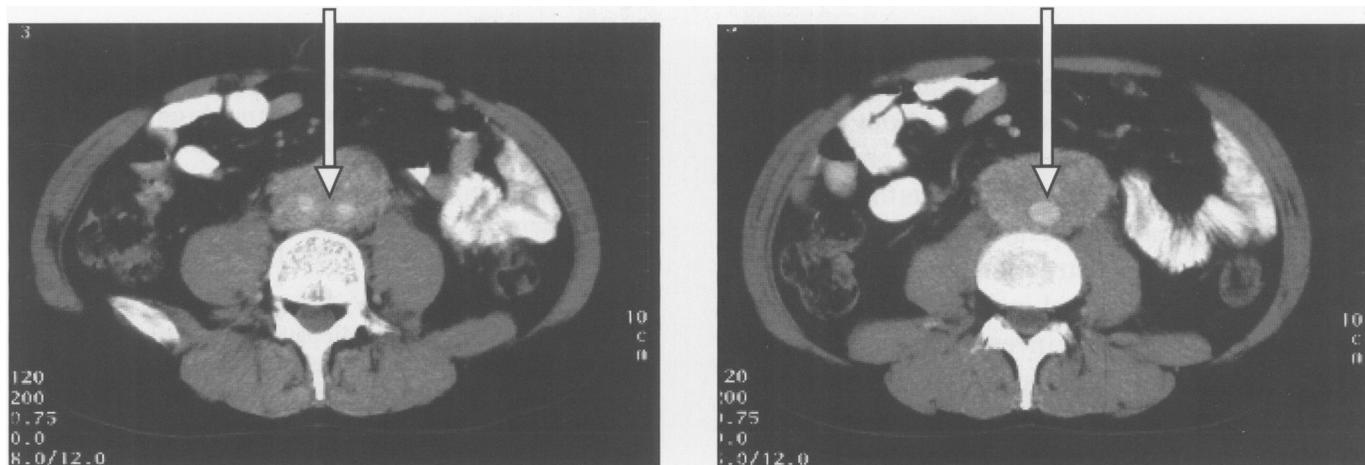


Fig 2A and 2B. CT scans showing significant soft tissue mass with IMA passing through the mass and compressing it.

next had a flexible sigmoidoscopy which revealed an inflamed friable sigmoid mucosa with contact bleeding precluding further advancement of the scope. He was, at this stage, treated with prednisolone enemas and mesalazine on the presumption that this was an inflammatory bowel condition. He was referred for a barium enema as an outpatient; this showed a normal distal descending and sigmoid colon (Fig 1). However, pathology results of biopsies from the sigmoidoscopy, returned later, should show features consistent with ischaemic colitis.

He subsequently re-presented two more times with LIF pain but additionally had testicular swellings which were revealed by USS to be bilateral hydroceles. Because of the recurrent nature of his symptoms he was further investigated with a CT scan of the abdomen which revealed a significant peri-aortic soft tissue mass. The IMA was noted to be compressed by this mass (Fig 2A and 2B). By

this stage his diarrhoea had settled and his colon had a normal appearance on CT. A subsequent CT-guided biopsy (Fig 3) confirmed histological features of RPF. The kidneys were not obstructed. An intravenous urogram showed mild bilateral hydronephrosis only.

TREATMENT AND FOLLOW-UP

On confirmation of the diagnosis of RPF he was started on an initial oral dose of 60mg of prednisolone daily with remarkable symptomatic improvement: he has had no further abdominal pain or bloody diarrhoea. Follow-up CT abdomen done a year later showed only minimal peri-aortic fibrotic changes (Fig 4). His steroids have been reduced to 10mg daily sufficient to hold his CRP at or close to normal.

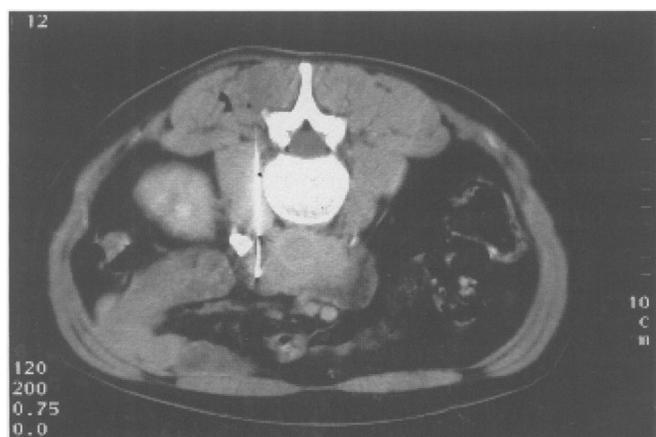


Fig 3. CT-guided biopsy with needle in mass.

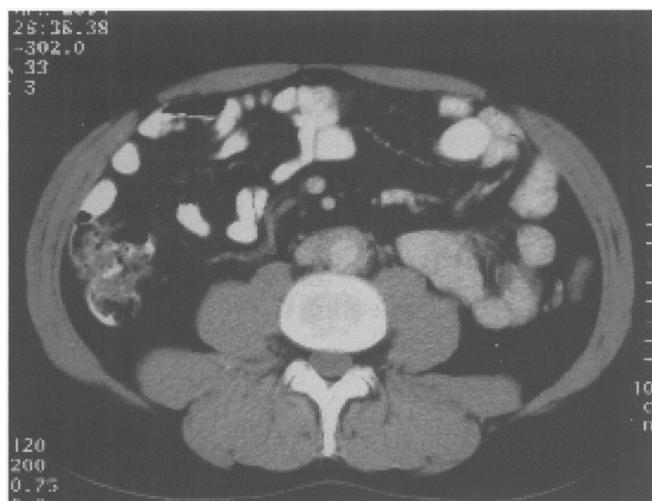


Fig 4. CT scan one year post treatment with steroids, at a similar level to fig 2, showing significant improvement.

DISCUSSION

RPF is a rare condition of uncertain aetiology but thought to be immune mediated. About two thirds of cases have no predisposing factors and hence are idiopathic. It characteristically affects the peri-aortic tissues often spreading laterally to involve the ureters leading to ureteric obstruction; the aorta and IVC are not usually displaced in RPF.¹ CT scan of the abdomen is the imaging investigation of choice.¹ It is important for the diagnosis to be confirmed either by radiological-guided or open surgical biopsy before commencement of medical treatment.⁶

Confirmed cases, even those with ureteric obstruction, can be successfully treated with steroids alone,⁶ and occasionally with tamoxifen,⁷ but ureterolysis may still be necessary in advanced or unresponsive cases.

This patient presented with non-specific abdominal pain later associated with bloody diarrhoea which was initially thought to be an inflammatory bowel disease from appearance of the large bowel on flexible sigmoidoscopy and the persistently high CRP and ESR. Biopsy however confirmed bowel ischaemia. Because he had further recurrences of this pain a CT scan of the abdomen was requested which then showed the mass, later confirmed to be RPF.

Although it is known that RPF usually encases the aorta, it is rare for this to significantly affect a major gut branch of the aorta and cause clinical ischaemia; there have been four reported cases worldwide^{2,3,4} but none, to our knowledge, has been reported in previous UK literature. Although Duffy TP did report a case of RPF presenting with left hydrocele⁵ this appears to be the first reported case presenting with bilateral hydroceles. This may be explained by the lymphatic drainage of the testis into the para-aortic nodes.

We suggest that RPF should be thought as a diagnosis in young persons presenting with bloody diarrhoea but with predominantly ischaemic changes in pathological biopsies.

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